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EDITORIAL: THE ONE AND A HALF VENTRICLE REPAIR—WE *CAN* DO IT, BUT *SHOULD* WE DO IT?

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In congenital heart disease, defects that are surgically septated are said to undergo a "two ventricle repair," whereas defects that cannot be easily septated surgically commonly undergo the Fontan operation, or "one ventricle repair." Occasionally, hearts have the internal morphology that allows surgical septation, that is, two ventricular cavities and two atrioventricular valves; however, the *morphologic and physiologic* characteristics of the right side of the heart are thought to be insufficient to carry a complete cardiac output. In recent years, the tendency has been to subject such patients to the so-called "one and a half ventricle repair." In this procedure the heart is surgically septated. This might include one or more of the following intracardiac procedures: atrial septal defect closure, ventricular septal defect closure, atrioventricular canal repair, a Rastelli-type left ventricular–aortic valve baffle with right ventricular–pulmonary artery conduit, and others. Additionally, all systemic–pulmonary artery connections are removed, and a superior cavopulmonary shunt is created. This latter component of the procedure eliminates all systemic-pulmonary mixing and provides preload reduction for the limited right heart, thereby avoiding right heart failure. Alternative procedures to the one and a half ventricle repair do exist for providing right heart preload reduction, for example, maintenance of an atrial septal defect or patent foramen ovale; however, this option achieves this goal at the expense of obligatory right-to-left shunting and cyanosis.

Patient selection for the one and a half ventricle repair comes under consideration in two specific clinical border zones. In one, the patient is a borderline candidate for a two ventricle repair, and the surgeon "backs off" to a one and a half ventricle repair to avoid postoperative right heart failure. Examples might include the patient with pulmonary atresia and intact ventricular septum with a right ventricle that is reasonably well developed but demonstrates *concerning morphologic and/or physiologic characteristics,* or the patient with Ebstein's anomaly and a tricuspid valve apparatus and/or right ventricular cavity, which demonstrates other equally *concerning morphologic and/or physiologic characteristics.*

At the other clinical border zone, the patient is clearly not a candidate for a two ventricle repair, and the focus over the course of the patient's life has been toward

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achieving a Fontan operation; however, the internal morphology of the heart does allow a surgical septation. Such patients are likely to have previously undergone palliative surgery, typically a neonatal procedure followed by a later bidirectional superior cavopulmonary shunt. Examples include the patient with a small right ventricle, inlet ventricular septal defect, and straddling tricuspid valve or the patient with D-transposition of the great vessels, conoventricular septal defect, and small right ventricle and tricuspid valve. The surgeon assesses the "difficulty factor" in achieving the intracardiac septation, as well as the size and function of the small right heart, and then makes a decision whether or not to proceed with the one and a half ventricle repair. The implicit assumption in this line of thinking is that avoiding the Fontan procedure is beneficial.

In both of these clinical circumstances the surgeon opts for the one and a half ventricle repair because it is believed that this procedure will result in improved functional status for the patient relative to the alternative operation. Can this view be substantiated? Certainly, many surgeons have clear anecdotal cases in which attempted two ventricle repairs failed decisively in the operating room after cardiopulmonary bypass or shortly thereafter, and the patients were salvaged quite effectively by a superior cavopulmonary shunt. Such cases represent the extreme and tell us very little in general about the right heart characteristics that will result in a beneficial outcome when the one and a half ventricle repair is used. When the one and a half ventricle repair is chosen over the Fontan procedure, even anecdotal evidence is lacking that the patient benefits.

What do we really know about the one and a half ventricle repair? We certainly know that the operation can be accomplished with acceptable surgical morbidity and midterm functional results. A review of the literature on the topic,[1-6] including the manuscript published in this month's Journal by Kreutzer and associates, tells us only that the procedure can be performed relatively safely and that patients seem to do reasonably well at follow-up.

What do we not know about the one and a half ventricle repair? At the end of the clinical spectrum that involves a decision between a two ventricle repair and a one and a half ventricle repair, we do not know what the specific criteria should be for choosing between the two operations, and we do not know whether individuals with right hearts that have *concerning morphologic and/or physiologic characteristics* actually are more functional with one and a half ventricle repairs or two ventricle repairs. Readers may object that the term *concerning morphologic and/or physiologic characteristics* used in the previous sentence and elsewhere in this discourse is too vague to have any real quantifiable meaning. They are correct. However, this is the extent of our knowledge in this regard. Nothing in the literature even remotely addresses the issue of making a quantitative assessment of the right side of the heart that can be used to objectively decide between the surgical options. The surgeon is left to make a clinical (subjective) decision between the two operations. The literature tells us we can successfully perform the operation, but it does not tell us who the candidates should be.

At the other end of the clinical spectrum, we also do not know whether the one and a half ventricle approach results in any benefit over that of the Fontan procedure. Even if we were to assume that the one and a half ventricle repair did provide some functional benefit in specific cases, no data are available to allow us to make a rational decision as to when the right heart characteristics (size? function?) reach the point at which the benefit is achieved in avoiding the Fontan operation.

We have known for a while that the one and a half ventricle repair can be performed safely. The studies that document this point represent important contributions. The safety of the procedure and the relative well-being of patients at follow-up have been established. Future publications making this point alone will add nothing to our understanding of the problem. We still must learn, however, whether performing this procedure is ever the right thing to do, both when the alternative option is the Fontan procedure and when it is the two ventricle repair. We need to define the *physiologic and morphologic criteria* that can be used in deciding on the optimal procedure for a given patient.

How can we acquire the information necessary to formulate these criteria? It is relatively easy to assess retrospectively a series of patients who have had one and a half ventricle repairs and show that these patients do reasonably well in follow-up. Acquiring the information that documents that the procedure is *beneficial* relative to the alternative operation (at both ends of the clinical spectrum) is more difficult, yet it is badly needed. One place to begin would be to define a large population of patients having one and a half ventricle repair and assign them to one of two categories based on whether the repair was performed as an alternative to a two ventricle repair or to a Fontan operation. The Fontan alternative category could then be compared to a group of actual Fontan patients. The comparison group for the two ventricle repair alternative category is more difficult to formulate; however, a cohort of patients with pulmonary atresia and intact ventricular septum who have undergone a two ventricle repair might be a reasonable

place to start. Various end points could be examined in these comparisons, including baseline functional status and exercise capacity. A careful quantitative evaluation of the preoperative morphologic and physiologic characteristics of the right side of the heart for all patients could then be used to determine whether, and under what right heart conditions, the one and a half ventricle repair is the more beneficial option. Such information would be welcome and noteworthy.

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